[CASE REPORT AND LITERATURE REVIEW]

Amyopathic Dermatomyositis Complicated by Pneumomediastinum

[a]RANDY TANG, RN, BSN, BSE, MD; [b]CHRISTIAN R. MILLETT, MD; [c]JUSTIN J. GREEN, MD

[a]UMDNJ—Robert Wood Johnson Medical School, Camden, New Jersey; [b]Dermatology Center, Bethesda, Maryland; [c]Cooper University Hospital, Camden, New Jersey

ABSTRACT

Dermatomyositis is an inflammatory disease of unclear etiology with characteristic cutaneous and musculoskeletal findings. Amyopathic dermatomyositis is a subtype without musculoskeletal involvement. Many cases of dermatomyositis are associated with underlying malignancy, but pulmonary manifestations can also be seen, the most common of which is interstitial lung disease. Pneumomediastinum is a rare complication that is important for clinicians to recognize, as it may be fatal if left untreated. The sudden onset of facial edema and shortness of breath in the setting of dermatomyositis should raise the suspicion of this condition. (*J Clin Aesthet Dermatol.* 2013;6(3):40–43.)

A 52-year-old Caucasian woman presented to the authors' clinic with a six-month history of fatigue, hair loss, and joint stiffness of the ankles, wrists, and knees. On physical exam, the patient was noted to have periorbital erythema with profound edema (Figure 1). Confluent and violaceous erythema was noted over the upper chest, and violaceous, ulcerated plaques were present on the metacarpalphalangeal joints, distal interphalangeal joints, and elbows bilaterally (Figure 2). A punch biopsy of the left elbow plaque was performed at the time.

The patient was started on prednisone 60mg once daily for a presumptive diagnosis of dermatomyositis (DM), and a comprehensive laboratory evaluation was ordered. The biopsy results were consistent with DM, and anti-Jo-1 antibody levels were normal. As her creatine kinase and aldolase levels were also normal, a diagnosis of amyopathic dermatomyositis (ADM) was made. At the next office visit, the patient showed no signs of clinical improvement, so hydroxychloroquine 200mg twice daily and clobetasol ointment were added. This combination led to significant improvement in the patient's cutaneous lesions.

One year after initial presentation, the patient came to the authors' clinic with an acute episode of shortness of breath as well as facial and neck edema. Crepitus was appreciated on physical exam. She was sent to the nearest emergency room where a computed tomography scan revealed bilateral pneumomediastinum (PnM) with subcutaneous emphysema. Bilateral chest tubes were placed at the time and cyclosporine therapy was initiated. The tubes were removed and the patient was discharged one week later after significant improvement in her condition.

DISCUSSION

Dermatomyositis is an inflammatory disease of unclear etiology characterized by proximal muscle weakness and cutaneous manifestations, including erythema of the eyelids with periorbital edema (heliotrope rash), erythematous to violaceous papules over the extensor aspects of the interphalangeal joints (Gottron's papules) as well as elbows and knees (Gottron's sign), hyperpigmentation with hyperkeratosis and fissuring of the hands (mechanic's hands), and erythema of the neck and upper chest in a v-shaped distribution (shawl sign). Amyopathic dermatomyositis is a subtype of DM in which patients present with typical cutaneous findings without associated muscle weakness or elevated muscle enzyme levels. The etiology of DM is a vasculopathic process with deposition of immune complexes on the endothelium of the capillaries in the skin as well as in the muscle.[1,2] Severe cases of DM may therefore present with ulcerative or even necrotic skin lesions.

It is well established that DM is associated with malignancy.[3,4] In some instances, the cutaneous

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ADDRESS CORRESPONDENCE TO: Christian Millett, MD, Dermatology Center, 6410 Rockledge Drive, Suite 201, Bethesda, MD 20817; E-mail: millett.christian@gmail.com

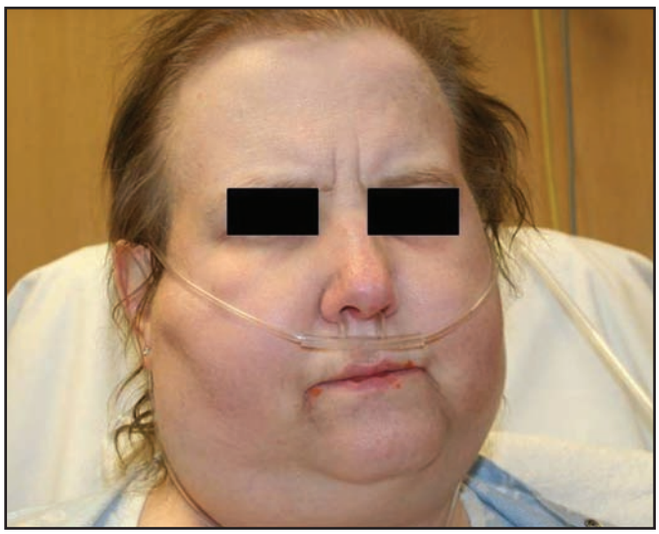

Figure 1. Significant facial and neck swelling of acute onset

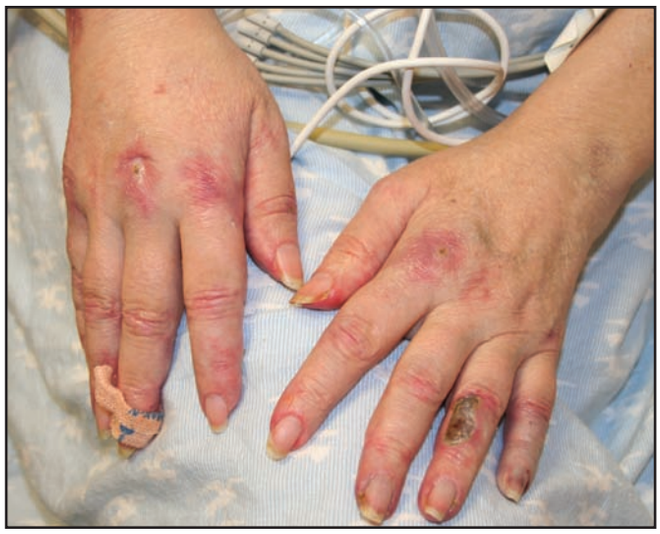

Figure 2. Erythematous, ulcerated papules overlying the MCP, PIP, and DIP joints of the hands

manifestations of DM are the first indication of an underlying malignancy. Additionally, it is estimated that approximately 50 percent of patients with DM will develop some type of pulmonary manifestation of their disease.[5] These include interstitial lung disease (ILD), aspiration pneumonia from esophageal dysfunction, pulmonary edema secondary to DM-induced cardiomyopathy, infectious pneumonia from drug-induced immunosuppression, pulmonary fibrosis secondary to methotrexate or cyclophosphamide therapy, or hypoventilation secondary to inflammation of the respiratory muscles. The most common pulmonary complication of DM is interstitial lung disease, and nonspecific interstitial pneumonitis (NSIP) accounts for 80 percent of these cases.[6] Patients with DM-associated ILD are more likely to have the anti-aminoacyl transfer RNA synthetase antibody (Anti-Jo-1), which has been termed the anti-synthetase syndrome. This antibody is present in 70 percent of DM patients with ILD, but only 20 percent of those without ILD.[7–9] However, the antibody is typically absent in patients with ADM-associated ILD.[10] Morganroth et al[11] reported that there was no difference in ILD prevalence between DM and ADM. Mukae et al[12] compared patients with DM-associated ILD and those with ADM-associated ILD and found a 45-percent mortality rate among those with DM as compared to a six-percent mortality rate for those with ADM.

Pneumomediastinum is a rare complication of DM that can be fatal if left unrecognized. Bradley[13] described the first case of PnM in an adult patient with DM, and since then, there have been several dozen reported cases, some in association with DM, others in association with ADM. Several mechanisms have been proposed to account for the origin of PnM in DM patients. One hypothesis is that PnM occurs when raised intra-alveolar pressure ruptures previously damaged alveoli in the lung or ruptures a subpleural cyst that developed from interstitial fibrosis.[14] A second hypothesis presumes that underlying pulmonary vasculitis leads to rupture of airway lesions,[15,16] while a third suggests that corticosteroids in the course of DM treatment weakens alveolar walls, predisposing to rupture.[17–19] Each of the above hypothesized mechanisms may contribute or act synergistically to increase the risk of PnM formation. The risk factors that predispose patients with DM to develop PnM therefore include the presence of interstitial lung disease (ILD), cutaneous vasculopathy, and a history of previous systemic corticosteroid treatment. Interestingly, younger age and absent creatine kinase elevation are risk factors as well.[20]

The treatments of choice for PnM are immunosuppressive agents, such as cyclosporine,[16,21,22,27] methotrexate,[17,18] and cyclophosphamide.[23] These should also be first-line treatments in patients with underlying ADM-associated ILD.[24] Other therapeutic options include the use of corticosteroids[25] and rituximab.[26] A review of previous cases highlights the variety of therapies that have been utilized (Table 1).

Although PnM may be fatal, many cases will self-resolve. Those cases reported in the literature that had a fatal outcome have typically occurred in individuals with concurrent ILD. The ultimate cause of death in these cases may not be from the PnM itself, but rather from respiratory failure secondary to progressive ILD. Hence, Yoshida et al[17] suggest that PnM in DM is not a prognostic factor. Rather, the presence and severity of ILD dictates the prognosis.[17,27]

Although uncommon, many physicians encounter patients with dermatomyositis. The sudden onset of facial edema and shortness of breath in the setting of DM should raise the suspicion of pneumomediastinum, as prompt recognition may be lifesaving. Other diagnoses in the differential include an allergic reaction, anaphylaxis, angioedema, and superior vena cava (SVC) syndrome.

TABLE 1. Previous cases and the variety of therapies that have been utilized

FIRST AUTHOR	AGE OF PATIENT/ SEX	MUSCLE SYMPTOMS	CREATINE KINASE	ILD	SKIN ULCERS	TREATMENT BEFORE SPnM	TREATMENT AFTER PnM	TIME TO RESOLUTION OF SPnM	OUTCOME
Powell[6]	34/M	No	Normal	Yes	No	PNS + AZA	Pulse IV methyl-PNSL 2g, PNS 80mg/day	N/A	Deceased
Bradley[13]	42/M	Yes	Elevated	Yes	Yes	PNS 80mg/day + AZA 100mg/day	Same as before	1 month	Alive
Yoshida[17]	38/M	Yes	Elevated	No	No	PNSL 40mg/day	3 weekly pulses of CPA 50mg +CSPN 300mg + PNSL 50mg/day + MTX 7.5mg/week	3 months	Alive
de Souza Neves[18]	45/M	Yes	Normal	Yes	Yes	none	PNSL 1mg/kg + chloroquine diphosphate 250mg/day + MTX 20mg +IV gamma globulins + pulse CPA 1g	2 months	Alive
Korkmaz[20]	28/M	Yes	Normal	Yes	No	PNSL 60mg/day	IV CPA 1g/month + PNSL 50mg/day tapered to 10mg/day after 4 weeks	3 months	Alive
Kim[21]	38/F	Yes	Elevated	Yes	Yes	PNSL 30mg/day + hydroxy-chloroquine 300mg/day	PNSL 30mg/day + hydroxychloroquine 300mg/day + CSPNA 100mg/day	1 month	Alive
Kuroda[22]	46/M	Yes	Elevated	Yes	No	PNSL 60mg/day	PNSL taper + CSPNA 100mg/day	4 months	Alive
Barvaux[23]	42/M	Yes	N/A	Yes	No	PNSL 0.5mg/kg + MTX 15mg/week + hydroxy-chloroquine 400mg/day	5 monthly IV pulses of CPA (0.75 to 1.5g/m^2) + IV gamma globulins (0.4g/kg for 5 consecutive days + single dose every 6 weeks	12 months +	Alive
Marsrouha[25]	66/M	N/A	Normal	Yes	No	PNS + MTX	Pulse steroid + MTX	4 months	Alive
Lee[26]	25/F	Yes	Elevated	No	No	PNSL 50mg/day + MTX 10mg/week	1g IV rituximab on Days 1 and 14, 740mg IV CPA + 500mg IV methyl-PNSL on Days 2 and 15	Several weeks	Alive
Terao[27]	16/M	Yes	Normal	Yes	Yes	Topical steroids	Bed rest, followed by PNSL 40mg/day + CSPNA 125mg/day	2 months	Alive
Current case	52/F	No	Normal	Yes	Yes	PNS 60mg/day + hydroxy-chloroquine 200mg twice/day	CSPNA 100mg twice/day, hydroxychloroquine 200mg/twice per day, PNS 30mg/day	1 week	Alive

ILD=interstitial lung disease; PnM=pneumomediastinum; PNS=prednisone; PNSL=prednisolone; AZA=azathioprine; CSPNA=cyclosporine A; CPA=cyclophosphamide; MTX=methotrexate, N/A=not available

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